Cancer Registry Issues in Studying Rare Cancers: A NAACCR Perspective

By

Holly L. Howe, PhD

Executive Director, NAACCR

Presented at

2nd NCI Epi Leadership Workshop

Understudied Rare Cancers

September 11, 2005

NAACCR

Definition is Big Issue

- Rare organ/histology combination
- Rare subtypes
  - Inflammatory breast cancer
  - Adenocarcinoma of the Lung
  - Medullary Thyroid Cancer
- Exposure-related Rare Cancers
  - Mesothelioma
  - Radiation-induced cancers

NAACCR

When in Doubt: Google

- Office of Rare Diseases – June 17, 2005
- ORD Search results were:
  - 85 Cancer sites listed
  - Site
  - Age (childhood)
  - Site/histology mix

ORD Listing since July 2005

- Anal cancer
- Endometrial cancer
- Fallopian tube cancer
- Lynch cancer family syndrome II
- Oropharyngeal cancer, adult
- Salivary gland cancer, adult
- Supraglottic laryngeal cancer

NAACCR

So again, what is a rare cancer?

- Orphan – no support, no home, no advocates, no population-based information
- Less than 200,000 cases
  - Time period? Place? Total in specific sub-populations?

In addition, Registry Issue is

- Validity of Diagnosis
  - Is it too rare to be plausible?
    - Reporting errors
    - Coding error
- Is it just very rare?
- How can we help Consortia?

CINA Deluxe, 1995-2002

- 7 provinces
  - Excludes 2 provinces; 2/3 country
- 42 states and D.C.
- Some states missing data for 1995-96
- 1,738,116,492 U.S. population (~60%)
- 67,075,774 Canadian population
- Data reported as of December 2004

NAACCR

Rare Childhood Tumors, 0-19 YEARS OLD, CINA, 1995-2002

	U.S.		Canada	
Bladder	Rate	Count	Rate	Count
All	0.56	314	0.48	9
Boy	0.71	202	0.52	5
Girl	0.41	112	0.45	4
Breast				
All	0.23	113	0.16	3
Boy	0.01	3	0	0
Girl	0.46	110	0.33	3
Colorectal				
All	0.81	398	0.37	7
Boy	0.8	202	0.21	2
Girl	0.82	196	0.55	5
Kidney				
All	6.69	3,356	6.74	118
Boy	6.12	1,572	5.84	52
Girl	7.29	1,784	7.68	66

	U.S.		Canada	
Larynx	Rate	Count	Rate	Coun
All	0.05	26	0.05	1
Boy	0.06	14	0.11	1
Girl	0.05	12	0	0
Rectum				
All	0.14	68	0.16	3
Boy	0.16	41	0.1	1
Girl	0.11	27	0.22	2
Sm Intestine				
All	0.09	42	0.11	2
Boy	0.08	21	0.1	1
Girl	0.09	21	0.11	1
Thyroid				
All	5.44	2,677	4.66	87
Boy	2.06	521	1.58	15
Girl	9.02	2,156	7.92	72

Rates are per 1,000,000 and age-adjusted to the 2000 US Std Population (19 age groups - Census P25-1130

Rare Childhood Tumors, 0-19 Yrs Old, CINA, 1995-2002

	United States				Canada				
Penis					**Testis**				
All	0.01	3	0	0	All	4.98	2,451	5.43	101
Boy	0.01	3	0	0	Boy	9.7	2,451	10.57	101
Pancreas					**Stomach**				
All	0.2	100	0.23	4	All	0.2	98	0.27	5
Boy	0.16	41	0.11	1	Boy	0.14	35	0.21	2
Girl	0.25	59	0.36	3	Girl	0.26	63	0.33	3
Oropharynx									
All	0.01	6	0	0					
Boy	0.02	4	0	0					
Girl	0.01	2	0	0					
Oral Cav/Pharynx									
All	2.24	1,109	1.93	36					
Boy	2.21	560	1.68	16					
Girl	2.28	549	2.21	20					

Rates are per 1,000,000 and age-adjusted to the 2000 US Std Population (19 age

Rare Tumors – NCI EGRP
Brain

	U.S.		Canada	
BRAIN	**Rate**	**Count**	**Rate**	**Count**
Male and female	**62.75**	**108,242**	**63.28**	**4,190**
Male	**74.93**	**59,672**	**76.15**	**2,408**
Female	**52.47**	**48,570**	**51.78**	**1,782**

Rates are per 1,000,000 and age-adjusted to the 2000 US Std Popn.

Rare Tumors – NCI EGRP
Endometrial, Ovarian, Testis

	U.S.		Canada	
CORPUS & NOS	**Rate**	**Count**	**Rate**	**Count**
Female	**241.61**	**225,996**	**216.26**	**7,517**
OVARY				
Female	**140.96**	**132,337**	**120.63**	**4,234**
TESTIS				
Male	**52.19**	**45,942**	**51.64**	**1,822**

Rates are per 1,000,000 and age-adjusted to the 2000 US Std Popn.

Rare Tumors – NCI EGRP
NHL, Myeloma, Kaposi Sarcoma, 1995-2002

	U.S.		Canada	
NHL	**Rate**	**Count**	**Rate**	**Count**
Male and female	**188.38**	**323,828**	**174.42**	**11,463**
Male	**226.27**	**171,492**	**206.94**	**6,249**
Female	**158.26**	**152,336**	**147.42**	**5,214**
MYELOMA				
Male and female	**54.15**	**92,958**	**49.53**	**3,215**
Male	**66.93**	**49,101**	**60.77**	**1,757**
Female	**44.89**	**43,857**	**40.66**	**1,458**
KAPOSI SARCOMA				
Male and female	**8.03**	**14,009**	**3.21**	**220**
Male	**15.13**	**12,679**	**5.78**	**194**
Female	**1.35**	**1,330**	**0.72**	**26**

Rates are per 1,000,000 and age-adjusted to the 2000 US Std Popn.

Rare Tumors – NCI EGRP
Liver, Stomach, Kidney

	U.S.		Canada	
Liver	Rate	Count	Rate	Count
Male and female	42.59	73,019	31.56	2,065
Male	66.83	50,822	50.4	1,526
Female	22.81	22,197	15.14	539
Stomach				
Male and female	77.69	133,382	90.35	5,854
Male	112.27	81,709	130.21	3,713
Female	51.93	51,673	59.03	2,141
Kidney				
Male and female	121.99	209,221	116.26	7,624
Male	167.6	127,638	154.72	4,684
Female	85.8	81,583	83.59	2,940

Rates are per 1,000,000 and age-adjusted to the 2000 US Std Popn.

Rare Tumors – NCI EGRP
Hodgkin Disease and Leukemias

	U.S.		Canada	
HODGKIN	**Rate**	**Count**	**Rate**	**Count**
Male and female	**27.78**	**48,550**	**25.14**	**1,719**
Male	**31.53**	**26,599**	**28.49**	**962**
Female	**24.44**	**21,951**	**22**	**757**
LEUKEMIAS				
Male and female	**121.04**	**208,450**	**122.61**	**7,984**
Male	**157.5**	**117,718**	**157.74**	**4,667**
Female	**94.13**	**90,732**	**94.42**	**3,317**

Rates are per 1,000,000 and age-adjusted to the 2000 US Std Popn.

Rare Tumors – NCI EGRP
Esophagus

	U.S.		Canada	
ESOPHAGEAL	**Rate**	**Count**	**Rate**	**Count**
Male and female	**49.24**	**84,493**	**40.81**	**2,654**
Male	**84.65**	**63,549**	**64.36**	**1,888**
Female	**21.28**	**20,944**	**21.13**	**766**

Rates are per 1,000,000 and age-adjusted to the 2000 US Std Popn.

Rare Tumors – NCI EGRP
Head, Neck, and Eye

	U.S.		Canada	
THYROID	**Rate**	**Count**	**Rate**	**Count**
Male and female	**69**	**119,307**	**56.74**	**3,866**
Male	**36.46**	**29,622**	**29.46**	**975**
Female	**100.42**	**89,685**	**83.6**	**2,891**
Oral Cavity and Pharynx				
Male and female	**108.52**	**185,814**	**100.72**	**6,632**
Male	**162.58**	**125,486**	**147.6**	**4,502**
Female	**63.52**	**60,328**	**60.31**	**2,130**
LARYNX				
Male and female	**45.42**	**77,867**	**30.99**	**2,037**
Male	**80.84**	**61,789**	**55.57**	**1,682**
Female	**17.24**	**16,078**	**10.18**	**355**
EYE				
Male and female	**8.25**	**14,201**	**8.13**	**531**
Male	**9.88**	**7,677**	**9.53**	**293**
Female	**7.01**	**6,524**	**6.87**	**238**

Rates are per 1,000,000 and age-adjusted to the 2000 US Std Popn.

Plans for Introductory Paper

- Define boundaries of “rare” since there doesn’t appear to be a standard
- Just provide statistics for really, really rare sites
- Descriptive epi statistics on those sites with sufficient numbers to subdivide among other characteristics:
  - Social/ Demographic
  - Geographic
  - Disease
- If fruitful, then convene interested research group to write a monograph.

Anyone interested in joining a rare tumor monograph group?

Contact me:
hhowe@naaccr.org

VISION for the Future

- Help consortia for rare tumor research by acting as a coordinating center [CC] for a RCA network
- Coordinate all registries to identify cases to CC
- QC diagnoses, obtain patient consents, secure bio-specimens
- Refer consented participants to investigators for interview

Beyond Surveillance

Many Advantages:

- **Large Number of cases**
- **Population-based research**
- **Training and consistency in RCA among expanded number of registries**
- **Efficiencies and consistency in RCA and triage**
- **RCA Network has been established in NAACCR (as of August 2005)**